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OCCLUSION OF THE ABDOMINAL AORTA AND DYSFUNCTION OF THE SPINAL CORD*

A CLINICAL SYNDROME

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SPONTANEOUS occlusion of the abdominal aorta as a clinical entity is usually accompanied by varying degrees of impaired blood supply to the lower extremities. The abnormal clinical features under these circumstances may be predominantly those of intermittent claudication, manifestations of peripheral neuropathy or ischemic necrosis of regional tissue. These syndromes may appear rather acutely or have a more prolonged progressive onset depending upon the nature of the occluding agent, whether embolus, thrombus or simply atheroma. Since the more obvious features are related to vascular embarrassment in the lower limbs, except in spontaneous dissection of the aorta, rarely has attention been directed to dysfunction of the spinal cord. It is our purpose to present another clinical syndrome, that of dysfunction of the lower spinal cord associated with occlusion of the distal aorta but without

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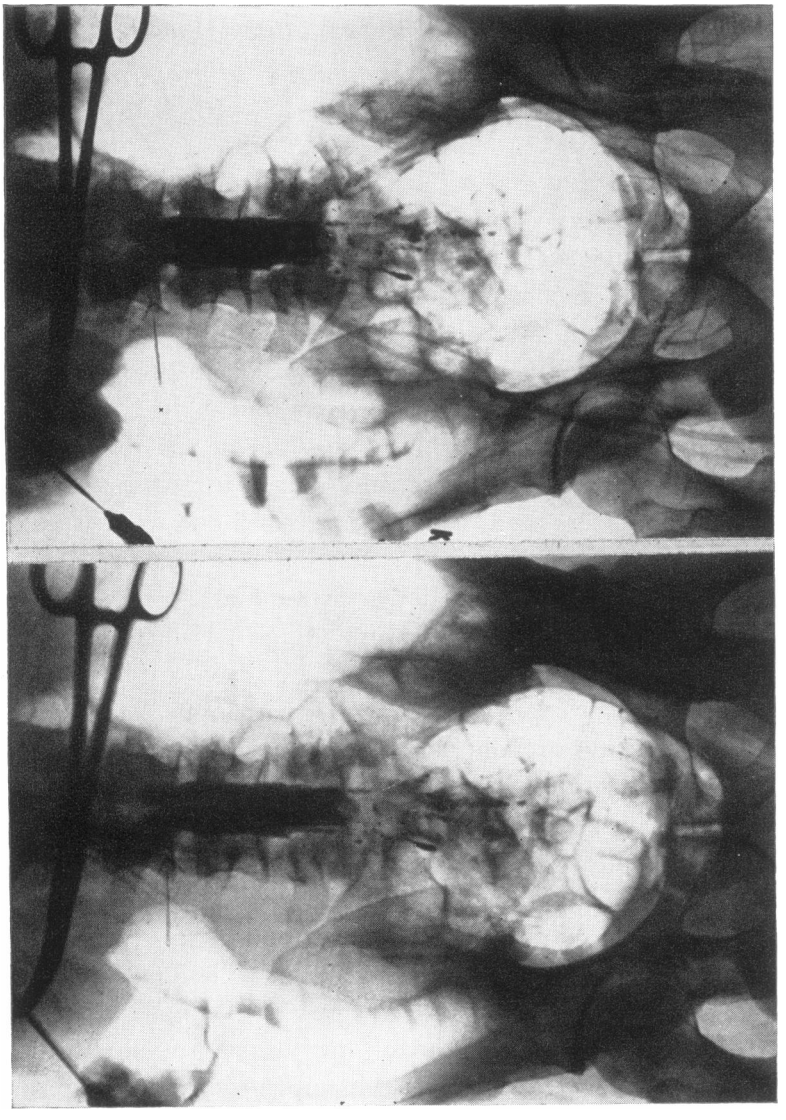
marked clinical impairment of circulation in the lower limbs. The essential features of the syndrome are as follows: There is rather rapid onset of varying degrees of paraparesis, the most cephalad part of the spinal cord implicated being from the seventh to the ninth thoracic segment. Significant alteration in appreciation of pin prick is present below this level and may be more evident on one side. Proprioceptive sensibility may or may not be disturbed. Areflexia or hyporeflexia is often present in the lower extremities and may persist. Bilateral Babinski signs are present in addition to incompetent vesicle and anal sphincters. Considerable pain of a radicular nature is most often present in the lower extremities. Plane roentgenograms of the spine do not disclose any abnormality, and the spinal subarachnoid space is found to be patent after a Queckenstedt test and myelography. Chemical and cellular examinations of the spinal fluid usually show normal results. After some time in the course of the illness, exploratory laminectomy has been done without disclosing the nature of the spinal cord lesion. Gradually, over a period of time, the level of altered sensibility regresses and may disappear. Muscle reflexes in the lower extremities may return to normal, yet areflexia may persist. Occasionally hyperreflexia and increased tone of the musculature of the lower extremities become evident. Babinski signs may also vanish; however, significant improvement in voluntary motor function of the lower extremities rarely occurs, particularly if the loss at the onset of the illness was severe in character. The degree and manifestations of recovery are variable in any single situation, and, in fact, evidence of return of spinal cord function may never become apparent. Confronted with this situation, the clinician often affixes the label of anterior spinal artery thrombosis to the syndrome, or the vague term "myelitis" is used to designate the clinical state. Suspicion regarding the underlying vascular nature of the lesion should be aroused by the course of clinical events enumerated. However, even though in some instances the circulation of the skin of the lower extremities appears adequate, one or more major peripheral pulses may be found to be absent or greatly diminished. In one patient the only suggestive vascular defect was an absent pulse in one femoral artery. In situations such as these, abdominal aortography will confirm the underlying vascular basis of the spinal cord disease by disclosing occlusion of the distal abdominal aorta. At the present time six such instances have been encountered.

ILLUSTRATIVE CASES

1. *Rheumatic Heart Disease, Auricular Fibrillation, Sudden Onset of Paralysis of the Lower Extremities with Evidence of Dysfunction of the Distal Spinal Cord. Absence of Gangrene. Absent Pulses in the Lower Extremities. Proven Aortic Occlusion by Aortogram and Operation.*

J. P., white male, aged 37 years, was admitted to the hospital because of the sudden onset of excruciating pain in the low back and lower extremities with "numbness and paralysis" of the lower limbs. During the preceding 15 years, recurrent difficulties related to rheumatic heart disease had been present. One year prior to entry into the hospital, transient hemiparesis had been evident on the right side for five hours. Nine days before hospitalization, following stretching in bed, severe lancinating pain appeared in the right lower quadrant of the abdomen and in the right flank. Cystoscopic examination did not disclose a suspected calculus in the urinary tract. Pain persisted until one week later when, following a bowel movement, excruciating pain occurred in the low back with radiation into both lower extremities. At this time the lower limbs could not be moved and were "numb". Urination was possible. Upon admission to the hospital one day later, pain was intense; the left lateral decubitus position was preferred, with hyperextension of the back. Attempts by the examiner to move the patient only evoked responses indicating severe pain, and the lower limbs were held flexed at the hips and knee joints. Rectal temperature was 100.6. The rhythm of the radial pulse was grossly irregular, with a rate of 120. The respiratory rate was 24. The arterial blood pressure was 140/90, while auscultation of the heart disclosed auricular fibrillation. The remaining abnormal pertinent physical features were confined to the lower limbs and trunk. Both lower extremities were warm. Good capillary and venous filling was demonstrated, even though the normal arterial pulsations of all vessels in both lower limbs were absent. In the right groin a cordlike structure, presumed to be a thrombosed femoral artery, was present, and abdominal aortic pulsations could not be felt below the midabdominal area. Marked spasticity was present in the lower extremities. The hamstring muscles were taut and voluntary motor function



A B

Fig. 1. Roentgenograms demonstrate pantopaque in the spinal subarachnoid space from myelography which did not disclose a lesion. Superimposed aortogram demonstrates (arrow) absence of filling of the distal abdominal aorta immediately after injection of opaque material and twenty seconds later. In the later roentgenogram (B) collateral filling of the vessels to the lower extremities is evident.

was absent. All muscle reflexes in the lower extremities were absent. The lower superficial abdominal reflexes and cremasteric reflexes were also absent, but minimal abdominal musculature responses were obtained after stroking the skin of the upper abdomen bilaterally. There was no response to plantar stimulation. Proprioceptive sensibility was absent in both lower extremities and appreciation of pin prick was markedly reduced in the right lower extremity up to the knee and on the left side to the groin. Analgesia was present over the entire posterior aspect of both lower limbs, but the saddle area and genitalia retained normal sensation. Roentgenograms of the chest disclosed a "mitral configuration" of the heart, and roentgenographic examination of the thoracolumbar spine did not disclose any abnormality. Calcification was not demonstrated in the abdominal aorta. The cellular content of the peripheral blood was normal. Urinalysis disclosed albuminuria. On lumbar puncture the spinal fluid was found to be clear and manifested no cellular or chemical abnormalities. Serological examination of blood and spinal fluid gave normal results. A spinal subarachnoid block could not be established by appropriate methods. In the following two days, pain decreased remarkably. "Pins and needles" appeared in both lower extremities and analgesic areas were now found to be hypalgesic. Proprioceptive sensibility remained markedly disturbed. Movement was beginning to return to the right leg. Three days later, the skin temperature of the right and left foot decreased and the distal parts of the lower limbs became pale; paralysis again returned to the right lower extremity. Appreciation of pin prick was now disturbed to just below the costal margin bilaterally. Superficial abdominal reflexes were not obtained and muscle reflexes were still absent in the lower extremities. There were no sphincteric difficulties. Gradually some voluntary movement returned to the right lower extremity and finally to the left lower extremity, the most marked paresis being present in the distal parts. An aortogram disclosed occlusion of the distal abdominal aorta (Figure 1, A and B). This finding was subsequently verified at operation, at which time a thrombo-endarterectomy (Figure 2) was performed. This procedure did not have any effect on the neurological syndrome. There was never evidence of gangrene. The last examination several months later disclosed some improved motor function in both lower extremities. Muscle reflexes in the lower extremities remained absent, but the superficial abdominal reflexes had returned to normal status in the upper quadrants of the

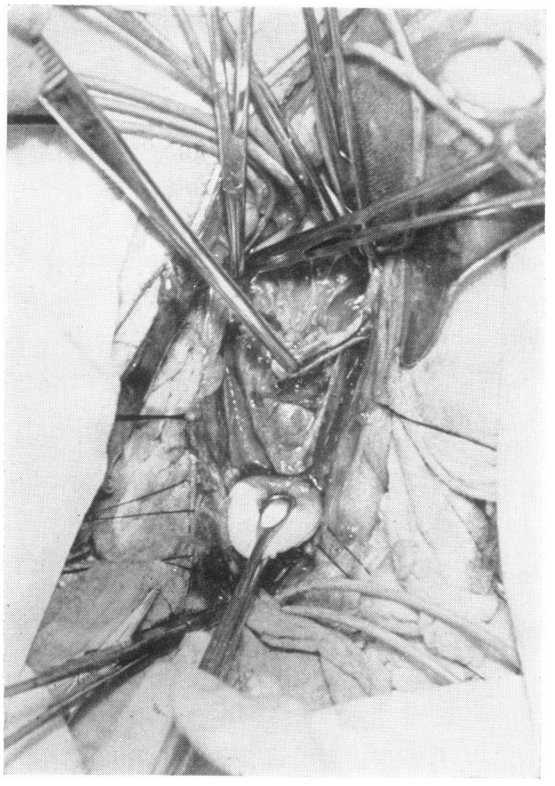


Fig. 2. Photograph taken at the time of thromboendarterectomy of the abdominal aorta (Case 1).

abdomen. Hypalgesia was still present to just below the costal margin, although normal sensation was preserved over the saddle area.

Comment

The neurological features in this patient indicated a lower motor neuron lesion as well as a tract lesion affecting the distal spinal cord. The subtending factor was lodgment of an embolus at the bifurcation of the distal abdominal aorta with impairment of circulation to the spinal cord, presumably through one of the radicular arteries. Temporary amelioration was followed by more cephalad extension of the abnormal neurological features. This may have resulted from propaga-

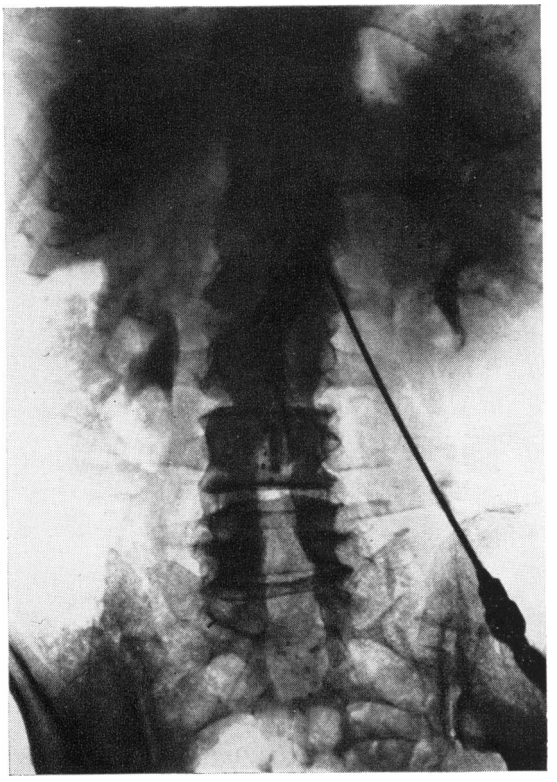


Fig. 3. Abdominal aortography indicating occlusion of the abdominal aorta (Case 2). Evidence of previous laminectomy is seen.

tion of a thrombus proximally. Of particular significance is the fact that occlusion of the distal abdominal aorta was manifested clinically solely by the neurological syndrome as outlined, without significant alteration of the skin circulation of the lower extremities.

2. *Sudden Onset of Bladder and Anal Sphincteric Incontinence. Mild Paraparesis. Laminectomy did not Disclose an Obvious Intradural Lesion. Gradual Amelioration of Neurological Symptoms in the Lower Limbs. Persistent Sphincteric Inadequacy. Aortogram Demonstrated Occlusion of the Distal Aorta.*

M. L., a middle-aged female, was able to recount the following history when examined some four years after the initial onset of her difficulty. She was in good health when she awoke one morning to find a puddle of urine in her bed. A physician was consulted immediately. It was observed at this time, however, that walking was somewhat difficult. Cystoscopy was performed and allegedly there was no reason disclosed for the urinary difficulty except a "weak bladder". Following cystoscopy incontinence of feces was observed. Neurological examination was performed at this time. The patient stated that there was numbness to pin prick up to the level of the midabdominal area. Walking was still possible but increasingly difficult. A myelogram failed to disclose evidence of a block, and a lumbar laminectomy also failed to disclose the etiology of the total bladder and anal sphincteric incontinence. Some two years later the problem was again reviewed neurologically. Hypalgesia was observed to the level of the knees bilaterally. Muscle reflexes were absent in the lower extremities. Approximately one year later, at the time of our examination, sphincteric incontinence remained total. There was vague hypalgesia over both legs and feet. The Babinski sign was present on the left but not on the right side. Motor power in both lower extremities was good. Both knee jerks were present. Ankle jerks were still absent. Proprioception was intact. Because of suspicion of aortic occlusion as the basis for this syndrome, aortogram was performed. This disclosed total obstruction of the abdominal aorta at the level of L_2-L_3 vertebrae (Figure 3).

Comment

The clinical history of rapid onset of symptoms referable to the spinal cord and the neurological pattern described by the patient with subsequent improvement aroused suspicion of a vascular lesion in the spinal cord. For this reason, the aortic lesion was disclosed by aortography.

Discussion

Vascular disease of the spinal cord occupies a curious position in neurological doctrine in that it is acknowledged almost only by the denial of its existence. Clinically, the sudden appearance of symptomatology and signs referable to the spinal cord is often considered in the vague category of "myelitis". Yet, if a clinical syndrome of the spinal

cord is classified under vascular disease, the anterior spinal artery is almost universally designated as the site of the disease process and the label "anterior spinal artery syndrome" firmly affixed. Acceptance of such notions has perhaps prevented the more frequent reporting of the syndrome described.

In 1911 Dejerine[1] described a condition which may be intimately related to the type of vascular disease of the spinal cord in the patients under consideration. Reichert, Rytand and Bruck[2] (1934), in fact, thought that such a syndrome was related to atherosclerosis. In their patients there was heaviness and weakness of both lower extremities, appearing and becoming intense during exercise, with relief following a period of rest. There were no signs of vascular insufficiency in the lower limbs, and the neurological abnormalities consisted of a mixed upper and lower motor neuron type of disorder. Mention was not made of the possible role of disease of the abdominal aorta at this time but it was felt by Dejerine that the basic disturbance was intermittent insufficiency of the circulation of the spinal cord. In 1925 Hunt and Cornwall[3] reported two instances of paraplegia associated with thrombosis of the distal abdominal aorta, and Hirsch[4] described an instance of myelomalacia associated with aortic occlusion. Apparently, these are the only verified reported instances of spontaneous occlusion of the abdominal aorta, manifested clinically primarily by a spinal cord syndrome, and without the so-called usual signs of vascular insufficiency in the lower limbs. In spontaneous dissection of the aorta, in which there are clinically recognizable signs of spinal cord dysfunction, the resultant clinical features are dependent upon the level of dissection in the aorta and the number of successive radicular arteries implicated. In our own experience, as well as that of others, instances have been encountered of dissection of the abdominal aorta where the significant late residua have been varying degrees of dysfunction of the spinal cord, but originally, vascular changes of fairly marked nature had been present in the lower limbs. This is in contrast to the patients considered here, in which there is little suggestion at all of the underlying vascular basis of the spinal cord dysfunction, except from the mode of onset and possibly the change in the peripheral pulses, but without significant circulatory embarrassment in the lower limbs. In the group of patients under consideration, the spinal cord dysfunction is always referable to the lower spinal cord segments and does not extend cephalad above the

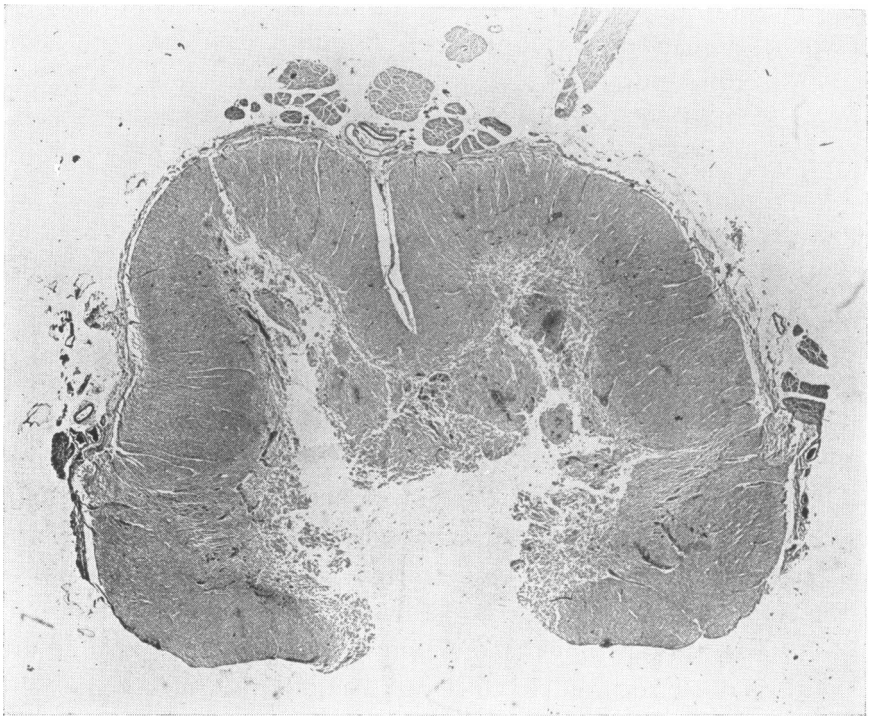


Fig. 4. Photomicrograph showing necrosis of the spinal cord of patient with occlusion of the abdominal aorta with the clinical syndrome of only dysfunction of the spinal cord.

eighth dermatomic zone. There is also evidence of lower motor neuron disease referable to these lower segments. The constancy of this pattern makes it evident that the radicular artery or arteries from the lower aorta are implicated. Aortography in these situations will often show the underlying vascular basis of the syndrome. It is of importance to note, however, that spontaneous occlusion of the distal abdominal aorta alone will not ordinarily result in the syndrome described. The demonstration of the occlusion of the abdominal aorta simply provides ground for speculation that a similar process has affected the segmental radicular arteries regionally as well as proximal to the occluding lesion, resulting in necrosis of the spinal cord (Figure 4).

Summary and Conclusions

A syndrome manifested primarily by dysfunction of the spinal cord but associated with occlusion of the abdominal aorta without the usual circulatory change in the lower extremities is described. The precise clinical characteristics observed warrant the utilization of aortography to establish the underlying vascular basis of the disease. Mention is made of other syndromes whose misinterpretation in the past has permitted the present entity to be generally unrecognized.

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